

Case report

Pregnancy with aortic dissection in Ehler–Danlos syndrome. Staged replacement of the total aorta (10-year follow-up)

Gérard Babatasi *, Massimo Massetti, Satar Bhooyroo, André Khayat

Department of Thoracic and Cardiovascular Surgery, University Hospital CHU Caen, Côte de Nacre, 14 033 Caen, France

Received 1 April 1997; received in revised form 16 June 1997; accepted 18 June 1997

Abstract

Pregnancy complicated by aortic dissection in patients with hereditary disorder of connective tissue presents interesting considerations including management of caesarean section with the unexpected need for cardiac surgery in emergency. Generalizations can be made on management principles with long-term follow-up requiring an aggressive individualized approach by a multidisciplinary team. A 33-year-old parturient presenting an aortic dissection at 37 weeks gestation required prompt diagnosis of Ehlers–Danlos syndrome in combination with correct surgical therapy resulted in the survival of both the mother and infant. During the 10-year follow-up, multiple complex dissection required transverse aortic arch and thoracoabdominal aortic replacement. © 1997 Elsevier Science B.V.

Keywords: Ehlers–Danlos syndrome; Marfan syndrome; Aortic dissection; Pregnancy

1. Introduction

It has been estimated that approximately 50% of all dissections in women less than 40 years of age occur during pregnancy or puerperum [1,2]. Ehlers–Danlos syndrome (EDS) is a genetically transmitted disorder of connective tissue characterized by hyperelasticity of the skin, hyperflexibility and looseness of the joints and arterial fragility with proneness to rupture [3,4]. It is now well documented that those patients have increased vascular complications [3], especially in the pregnant patient [4].

2. Case history

On march 1987, a 33-year-old woman remained well for her first pregnancy until the 37 weeks when she complained of acute thoracic pain radiating through her back. She was admitted to the gynecology outpatient department. Past medical history included surgical treatment for cyphoscoliosis, herniorrhaphy and bullous emphysema. She was hypertensive with a blood pressure of 166/70 mmHg. On auscultation, aortic insufficiency was detected. Chest X-ray revealed cardiac enlargement in the upper mediastinum and right pneumothorax. Trans-esophageal echocardiography (TEE) and CT scan were performed showing aortic insufficiency of grade II and a flap into the ascending aorta with a false lumen on the right side (acute type A

* Corresponding author. Tel.: +33 02 31064457; fax: +33 02 31064521.

dissection). Operation was planned as a cesarean section with subsequent aortic repair. Hypertension was controlled with hydralazine infusion. After transverse laparotomy, a healthy male child was delivered. While closure of the uterotomy and the abdomen was performed by the obstetricians, the right femoral artery was then cannulated for cardiopulmonary bypass (CPB). After sternotomy and distal aortic cross-clamping, the ascending aorta was opened showing the dilated false lumen, a 2-cm tear, distal to the noncoronary aortic cusp was detected as the proximal entry. Dissection was circular, requiring resuspension of the aortic valve. After cardioplegia, the ascending aorta was transected immediately proximal to the innominate artery. The false lumen was obliterated by sandwiching the two aortic wall layers between a Teflon felt strip. The aortic root was reconstructed using a 30 mm woven graft (Meadox Medicals, Oakland, NJ) sutured to the aortic annulus, using multiple everting interrupted sutures reinforced by Teflon felts. The graft was attached end-to-end to the distal ascending aorta. Bilateral blebs was resected using stapling device. Microscopic sections of the aorta revealed focal areas of collagen bundle disruption and an increase in elastin relative to collagen within the fibromuscular wall. A skin biopsy revealed a thin dermis with increased elastin consistent with the clinical diagnosis of EDS.

In June 1990, an infrarenal abdominal aortic aneurysm (55 mm diameter) due to retrograde left iliac artery reentry was detected on CT scan. An aorto-biiliac bypass graft using a 24 mm prosthesis was performed.

On May 1992, a dilatation of the transverse aortic arch of 8 cm in diameter, including the supraaortic vessels, was diagnosed on CT scan. Arch replacement under CBP, deep hypothermia and cold blood cerebroplegia was performed using the 'elephant trunk technique'. Reimplantation of the supra-aortic arteries was made in a 34 mm woven graft. The graft was inverted on itself and then inserted in the descending thoracic aorta. The inverted edges of the graft were sutured to the descending aorta past the left subclavian artery. Next, the inverted tube was retrieved in the mediastinum, and a side-hole was cut to which the brachiocephalic arteries were anastomosed.

At 8 months later, aortic graft replacement for treatment of thoracoabdominal complex dissection involving celiac, superior mesenteric and renal arteries was performed. Prostaglandine E₁ (100 µg/kg) was given intravenously to protect the cerebral microcirculation. Thoraco-abdominal step repair was performed by 'sequential clamping technique' under partial CPB. A 30-mm woven graft was inserted through a left thoracoabdominal incision. Visceral arteries were all reattached to the prosthesis using separate opening made in the graft (Fig. 1). The patient's postoperative course was uncomplicated.

3. Comment

Most of aortic dissections in pregnancy occur as a result of systematic hypertension [5]. Others risk factors included Marfan's syndrome, Ehler–Danlos syndrome (EDS), bicuspid aortic valve, coarctation of the aorta [6,7]. The outcome of pregnancy in patients with disorder of connective tissue and aortic dissection is not usually as favorable as that reported here [4,6]. The successful outcome achieved in our patient was undoubtedly influenced by many factors, including prompt diagnosis, preoperative hypotensive drug therapy, early concomitant cesarean section and cardiac surgery followed by annually serial CT scan imaging and echocardiographic studies of the whole aorta. There are several reports [1,7] of aortic dissection during pregnancy, although the rarity of this condition has resulted in no clear picture of the relative risk of dissection compared with the nonpregnant state. Estrogen has been reported to inhibit collagen and elastin deposition in the aorta, and progesterone has been shown to accelerate deposition of non collagenous proteins in the aortas of rat [8]. The increase in cardiac output with increased heart rate, left ventricular wall mass and stroke volume could increase shear stress in the aorta. Aortic dissection is essentially an accident occurring in a common process, that of continuous injury and repair as part of the ageing process, this is accelerated in Marfan or EDS because of the underlying connective tissue defect. Hydralazine and beta-blockers were recommended to control blood pressure and decrease left ventricular contractility without fetal toxicity [1].

Our report describe a patient with previously unrecognized Ehlers–Danlos that was first diagnosed during pregnancy. Up to eight varieties have been described [3]. Type IV, which was the type of our patient, is known as the arterial type and characterized by minimal joint laxity, very thin cigarette-paper-like skin, severe bruisability, and a marked tendency toward rupture of arteries [3,4]. Patients with type IV are prone to rupture of large arteries via a tear through the wall of the vessel rather than via dissection or trauma, and the fragility of the arterial wall makes surgical repair extremely difficult. Women with Marfan's syndrome or EDS who have minimal aortic root dilatation, aortic regurgitation are at a high risk of life-threatening cardiovascular complications during or shortly after pregnancy [1,4,9]. The safe option consist in cesarean section, if the fetus is viable, immediately before and at the same operation as the cardiac procedure [10]. The redo-procedures were imposed upon evolution towards aneurysms due to evolution of EDS type IV. All authors who have operated on EDS type IV patients describe the extreme friability of the vessels in this condition [3,4]. Sutures cut out, ligatures cut through

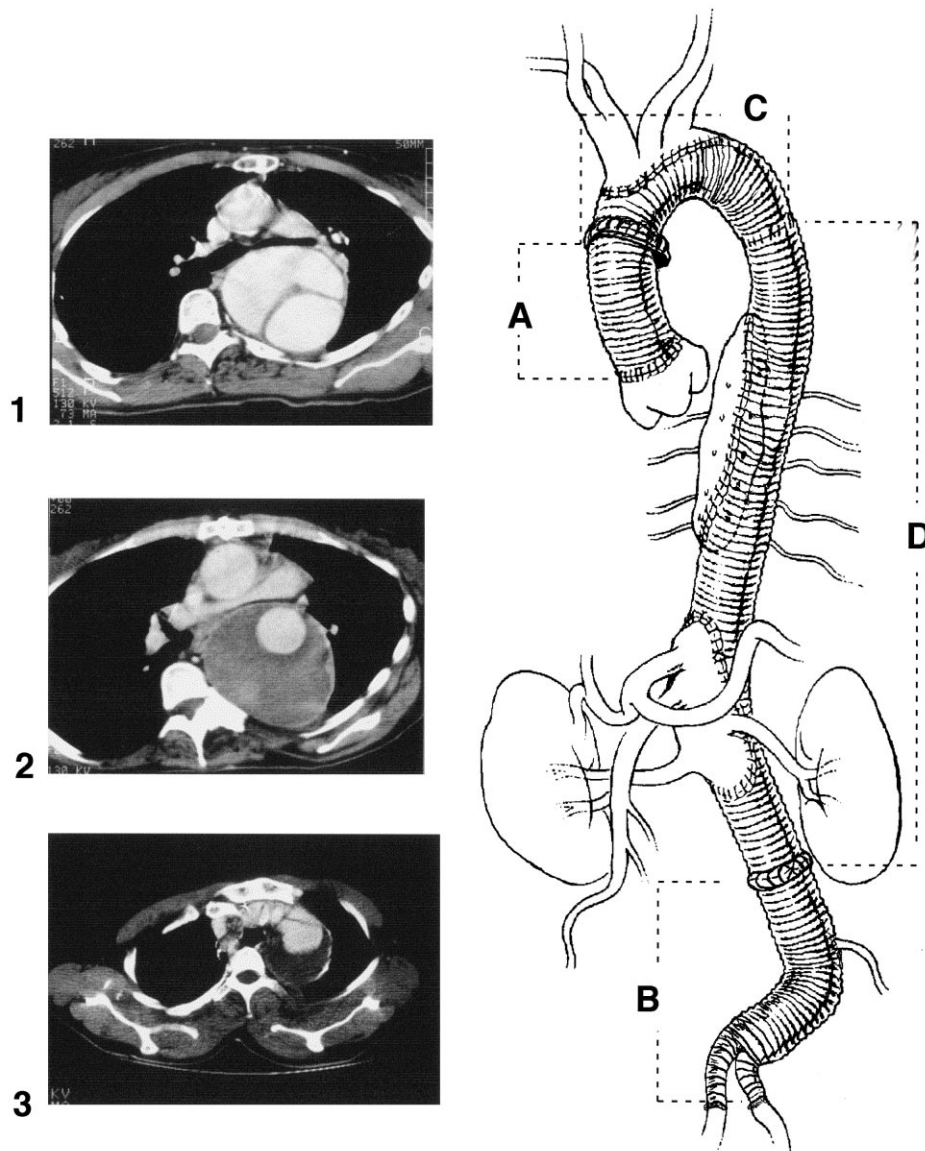


Fig. 1. Aortic multiple-stage procedures: (A) 1987—emergency surgery with concomitant cesarean section and bullous excision; (B) 1990— aorto-biiliac bypass graft; (C) 1992—transverse aortic arch replacement with the ‘elephant trunk technique’; (D) 1993—thoracoabdominal graft replacement with intercostal, celiac, superior mesenteric and renal arteries reattachment. On the left side, CT scan: 1: aortic Type I dissection 2: opacification of the Elephant Trunk into the descending aorta 3: aortic arch dissection.

and conventional arterial clamps may tear the vessels. In general, extensive surgical intervention in any form should be avoided if possible. After the emergency operation, CT scan was performed annually and detection of the 55-cm abdominal aortic aneurysm 3 years after the initial procedure was casually visualised. At that time, only moderate dilatation of the aortic arch was detected by angiography. After discussion with her cardiologist, medical control was annually planned. During a 2-year period, aortic arch reached 8 cm in size with extension to thoraco-abdominal aorta and indicated surgery.

The present case, as well as the others reported [3,4] suggested that patients with type IV of EDS should be

advised against pregnancy. If those patients elect pregnancy, she and her physician should at least be aware of the potential severe complications. Long term survival required a strict medical survey. A consensus is developing [11] that type III dissection in nonpregnant women are best treated by normalizing blood pressure, reserving surgical repair for the patients who remain symptomatic or have documented extension of the dissection, as seen in the follow-up of our patient. If replacement of the descending thoracic aorta with reattachment of intercostal or visceral arteries can be deferred for several months, sutures hold better and the procedure has less risk, especially in patients with EDS type IV. Transoesophageal echocardiogram, CT Scan

are at least annually required to improve the difficult management of such young patients.

Acknowledgements

We are indebted to Michel Cruet, Bruno Postel, Daniel Richard and Yves Dalibert for collaborative efforts during cardiopulmonary bypass, and to Jean-Louis Gerard MD, for anesthetic management.

References

- [1] Elkayam U, Ostrzega E, Shotan A, Mehra A. Cardiovascular problems in pregnant women with the Marfan syndrome. *Ann Intern Med* 1995;123:117–22.
- [2] Oskoui R, Lindsay J. Aortic dissection in women > 40 Years of age and the unimportance of pregnancy. *Am J Cardiol* 1994;73:821–3.
- [3] Wesley JR, Mahour GH, Woolley MM. Multiple surgical problems in two patients with Ehlers–Danlos syndrome. *Surgery* 1980;87:319–24.
- [4] Snyder RR, Gilstrap LC, Hauth JC. Ehlers–Danlos Syndrome and pregnancy. *Obstet Gynecol* 1983;61:649–50.
- [5] Pumphrey CW, Fay T, Weir I. Aortic dissection during pregnancy. *Br Heart J* 1986;55:106–10.
- [6] Konishi Y, Tatsuta N, Kumuda K, Minami K, Matsuda K, Yamasato A, Usui N, Muraguchi T, Hisaka Y, Okamoto E, Watanabe R. Dissecting aneurysm during pregnancy and the puerperium. *Jpn Circ J* 1980;44:726–33.
- [7] Anderson RA, Fineron PW. Aortic dissection in pregnancy: importance of pregnancy-induced changes in the vessel wall and bicuspid aortic valve in pathogenesis. *Br J Obst Gyn* 1994;101:1085–8.
- [8] Foidart JM, Rorive G, Nasyens B. Aortic collagen biosynthesis during renal hypertension, pregnancy and hypertension during pregnancy in rat. *Biomedicine* 1978;28:215–8.
- [9] Brearley S, Fowler J, Hamer JD. Two vascular complications of the Ehlers–Danlos syndrome. *Eur J Vasc Surg* 1993;7:210–3.
- [10] Parry AJ, Westaby SW. Cardiopulmonary bypass during pregnancy. *Ann Thorac Surg* 1996;61:1865–9.
- [11] Williams GM, Gott VL, Brawley RK, Schauble JF, Labs JD. Aortic disease associated with pregnancy. *J Vasc Surg* 1988;8:470–5.